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SPINAL CORD TUMOR IN A PATIENT WITH MULTIPLE SCLEROSIS

Case report

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ABSTRACT - The association between multiple (MS) sclerosis and cerebral gliomas has been sporadically reported in the literature, causing a long lasting discussion if these lesions occur coincidentally or if MS plaques may actually lead to the genesis of gliomas. We report a 36 year old man who developed a rapid onset of right side weakness and loss of vision, having established a diagnosis of MS which was confirmed by CSF analysis and MRI. Nine years later he developed progressive tetraparesis, leading initially to suspicion of illness relapse and a demyelinating plaque in the spinal cord. However, after MRI investigation, a spinal cord tumor was diagnosed. The patient underwent cervical spine laminotomy for microsurgical removal of the spinal cord tumor diagnosed as ependimoma. The neurological deficits improved significantly.

KEY WORDS: multiple sclerosis, gliomas, spinal cord tumors.

Tumor de medula espinal em paciente com esclerose múltipla: relato de caso

RESUMO - A associação entre esclerose múltipla (EM) e gliomas cerebrais foi relatada esporadicamente na literatura, levando a longa discussão quanto à possibilidade das placas de esclerose estarem envolvidas na etiologia dos gliomas ou dessas lesões ocorrerem coincidentemente. Relatamos um paciente de 36 anos que desenvolveu hemiparesia direita rapidamente progressiva e perda visual, sendo estabelecido o diagnóstico de EM após análise do LCR e imagens de RM de encéfalo. Após nove anos o paciente desenvolveu tetraparesia lentamente progressiva, levantando inicialmente a hipótese de atividade da doença e aparecimento de placa de EM na medula espinal. Contudo, após investigação com RM de coluna, um tumor medular foi diagnosticado. Foi então submetido a laminectomia cervical para ressecção microcirúrgica do tumor, que foi diagnosticado como ependimoma. Os déficits neurológicos melhoraram significativamente.

PALAVRAS-CHAVE: esclerose múltipla, gliomas, tumores medulares.

Multiple sclerosis (MS) is a demyelinating disease of the central nervous system (CNS) classically described as a relapsing-remitting disorder that affects multiple white-matter tracts, with usual onset in young adults and displaying marked clinical heterogeneity. The varied clinical features reflect the multifocal areas of CNS myelin destruction. The diagnosis of MS was traditionally based on clinical history, neurological examination and analysis of cerebrospinal fluid (CSF). Evoked potential examination and magnetic resonance imaging (MRI) have brought new aids to the diagnosis[1]. Frequently, a pattern of disseminated encephalomyelitis occurs, when several plaques of demyelinization can be observed throughout the neuroaxis. The plaques are typically surrounded by scar tissue, in which gliosis and glial cell division are observed. The differential diagnosis of MS is quite limited in the setting of a young adult with two or more clinically distinct episodes of CNS dysfunction. Difficulty arises when atypical presentations occur, such as monophasic episodes on a progressive illness. Great care must be taken to exclude treatable etiologies (compressive spinal cord lesions, arteriovenous malformations, cavernous angiomas, Chiari malformation).

The concomitance of MS and primary CNS tumors could be explained by simple coincidence based on the statistics of incidence and prevalence of both diseases. However, since few reports have shown the coexistenceof these conditions and it is not known how many patients actually carry them, such affirmation cannot yet be done. In the same way, it is

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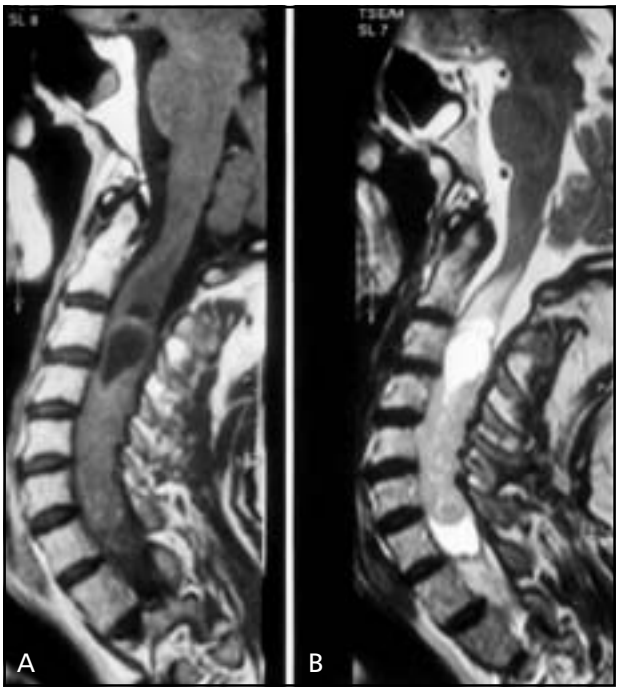

Fig 1a. T1 weighed sagittal MR with gadolinium contrast of the cervical spine exhibiting a large iso-signal-intensity mass lesion between C3 and C7, with a cyst in the upper level. Fig 1b. T2 weighed sagittal MR exhibiting a hyper-signal-intensity lesion and cyst.

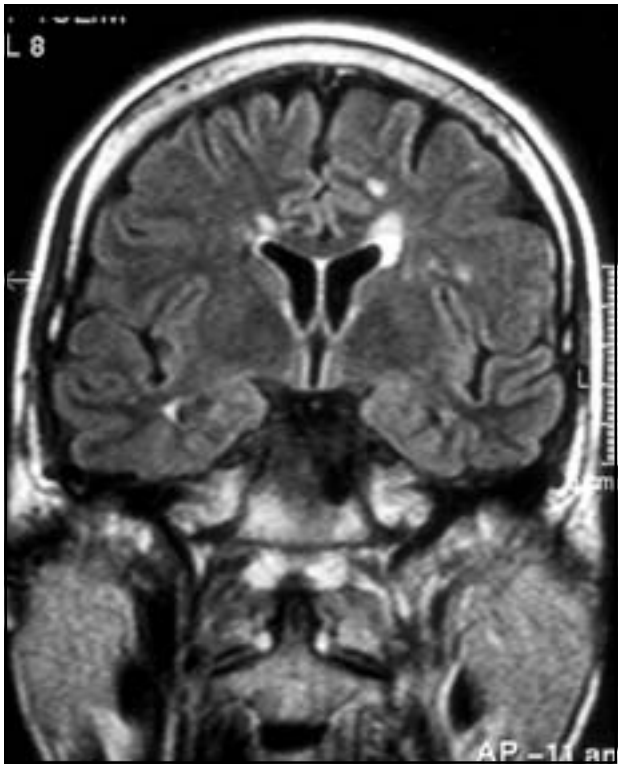

Fig 2. A coronal MR image exhibits multiple lesions in the white matter, suggestive of multiple sclerosis.

not possible to demonstrate that the gliosis around the MS plaques should be implicated in the genesis of the CNS tumors. Nevertheless, one could expect such transformation to be plausible, thinking from a strict pathophysiological point of view. There is a greater chance for a tumor to be initiated in a region of frequent mitosis than in another of infrequent mitosis. This risky area would be only for the initiation of gliomas, ruling out from this theory all other CNS tumors. Previous reports, including a review from Currie and Urich[2] have shown patients with clinical diagnosis of MS that developed cerebral gliomas, mostly malignant. We report on a case of a 36 year old man with a clinical diagnosis of MS who later on developed a spinal cord glioma.

CASE

A 36 year old man, business administrator, presented with a sudden onset of weakness and loss of sensitivity on the right side and loss of vision of the right visual field. It lasted for approximately four weeks and spontaneously improved to a lower than normal status. He underwent encephalic MRI which revealed white matter lesions suggestive of multiple sclerosis and a cerebrospinal fluid (CSF) tap was performed, exhibiting 1 leukocyte per ml and 37 mg of protein per 100 ml. The protein electrophoresis showed a peak of 17.3 % of gammaglobulin. Diagnosis of MS was established and he received initially prednisone therapy (40 mg daily) which was later changed to immunosuppressive treatment with azathioprine (100 mg daily) which was maintained for a total of seven years with a single one year interval.

Nine years after the first episode (when the patient was 45) he presented progressive spastic tetraparesis despite the use of regular immunosuppressive therapy. The medication was discontinued and a new MRI was performed, exhibiting a large spinal cord lesion with iso-intense-signal on T1 weighed MR (Fig 1) between the second and seventh cervical vertebrae, with a cystic nodule at the C3 level. At this time an encephalic T2 weighed MR exhibited small lesions suggestive of demyelinization plaques (Fig 2).

He was submitted to a C2-C7 laminotomy for microsurgical removal of the tumor. The pathological examination revealed an ependimoma. He recovered well, with gradual improvement of muscular strength. He now presents grade 3 to 4 strength on the left upper limb and grade 4 to 5 on the right. The lower limbs present grade 5 strength with exacerbated deep-tendon reflexes. The post operative T1 weighed spinal MR confirmed the complete resection and did not exhibit any surgical complication (Fig 3).

DISCUSSION

The concomitance of MS and cerebral gliomas has been reported by few authors in the literature. There has been questioning of a possible pathological re-

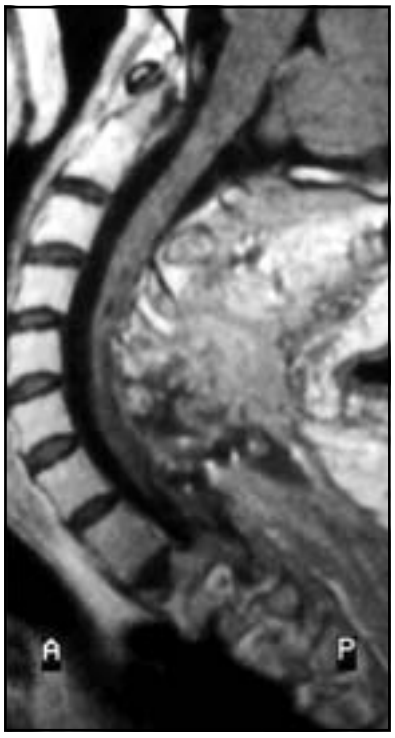

Fig 3. Post operative sagittal T1 weighed MR with gadolinium contrast of the cervical spine shows the removal of the lesion from the spinal cord, without evidence of surgical complications.

lationship between the two diseases, without conclusive evidence yet. Aarli et al.[3] have also proposed the possibility of the immunosuppressive treatment for MSfacilitating the initiation of primary CNS tumors. However, none of the cases reported in the literature received full immunosuppressive treatment, including that in Aarli's report. In the current report , the patient received a prolonged immunosuppressive therapy. Although there is no proof that lacking imunological defenses may have facilitated tumoral initiation, it is an important consideration to make when any tumor arises.

The possible tumor initiation from a MS plaque has been discussed mainly on a pathological basis. In the Currie and Ulrich[2] review, the main evidence of initiation comes from whether there is or not a plaque contiguous to the glioma on the microscopic examination of the specimen. It is questionable if the tumor was actually induced by the contiguous plaque or if it initiated elsewhere and extended itself to meet a "false" contiguous plaque. The same is valid for Brihaye et al.[4] and Lahl[5] cases. Nahser et al.[6] reported two cases in which no contiguity was observed. If the tumors developed from a plaque, it was possibly obliterated by the growing mass, making microscopic observation of contiguity very difficult. Russel and Rubinstein[7]also presented three cases with this association, one of which did not present any contiguity between the tumor and plaques, supposedly because the tumor cells destroyed the plaque as well.

Thus, whether gliomas occur in MS patients by mere coincidence or whether they are induced by the demyelinization plaques cannot yet be determined. A larger number of cases will need to be reported before MS becomes a known risk factor for the initiation of gliomas.Early investigation of MS patients with MRI may help in the future to determine if diagnosed plaques will give rise to tumors.

We present a report which is distinct from the others in two aspects. It is the first case of concomitant MS and primary spinal cord tumor, all others represented by the association of primary brain tumor and MS. Second, the diagnosis and report has been done with the patient alive, based on clinical and CNS imaging diagnosis. The small amount of spinal cord glial tissue (in comparison to the brain) and the rare occurrence of spinal cord plaques and tumors may explain why these lesions were not known to occur in concomitance.

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